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A valuable option in cases of severe mitral regurgitation and mildly enlarged aortic root in Marfan syndrome: the role of Personalised External Aortic Root Support.

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Abstract (99/100 words)

Two patients with Marfan syndrome are presented in whom there was mitral regurgitation meeting criteria for valve repair. Both patients had an aortic root aneurysm at the threshold of size criteria for prophylactic surgery and mild aortic valve regurgitation. Conventional mitral valve repair was performed. Personalised External Aortic Root Support (PEARS) was used to halt aortic root aneurysm progression. This also corrected the aortic regurgitation. Combining mitral repair with root support by PEARS avoided the dilemma of leaving the aortic root pathology uncorrected or engaging in more complex surgery than seemed justified at the clinical stage of the disease.

Introduction

Mitral valve prolapse is the second commonest cardiac manifestation of Marfan syndrome.

If patients with Marfan syndrome have mitral valve prolapse at the time of aortic root replacement, correction of mitral valve regurgitation is generally performed at the same time if the severity of the regurgitation is moderate or worse. If the indication for surgery is clinically important mitral regurgitation and there is an aortic root aneurysm just meeting size criterion of 4.5 cm [1] this presents additional operative complexity and risk. If not dealt with the aortic root remains prone to further dilatation and rupture.

Since 2004 personalised external aortic root support (PEARS), has been available as an alternative to valve sparing root replacement and has been under evaluation. A published summary of the current experience and results is provided in a follow up in the first 30 patients who are now on average 6.5 years after surgery.[5] PEARS is quite a different procedure from ‘wrapping’ the aortic root with stiff vascular graft material. The fabric used for PEARS is soft, pliable and macroporous. It becomes entirely incorporated to form a neo-aortic wall. The root size and shape are held fixed at the size of the customised mesh which is individually made using the data from the patient’s digital imaging. The mesh intimately covers the aorta from its junction with the left ventricle to beyond the brachiocephalic trunk.[3] Effectiveness of external support in correcting aortic valvar regurgitation has been shown with a vascular graft used similarly to a Florida sleeve.[2] It can also be achieved with the Personalised External Aortic Root Support (PEARS).[3;4]

PEARS has been available at a very few centres. The procedure provided a solution to the clinical problem in two patients with the combination of clinically important mitral

regurgitation and a root aneurysm at a size when guidelines indicate that it should not remain unoperated.

Cases

Case 1. A 17 year old man with diagnosis of MFS with severe mitral regurgitation due to posterior mitral leaflet prolapse also had a characteristic root aneurysm. The diameter at the level of apposition of the valve leaflets was 45 mm and there was mild aortic regurgitation. He was operated on in January 2015.

Case 2. A 55 year old woman with diagnosis of MFS with severe mitral regurgitation due to posterior mitral leaflet prolapse also had a characteristic root aneurysm. The aortic root diameter was also 45 mm and she too had mild aortic regurgitation. She was operated on in March 2015.

Intraoperative examination confirmed prolapse of P2 posterior mitral leaflet without chordal rupture in both cases. Both the mitral valve component and the PEARS operations are standardised procedures and were performed similarly in both patients. After establishing standard cardiopulmonary bypass, P2 resection and mitral annuloplasty was performed in each patient. The aortic cross-clamp was then released and a normal sinus rhythm was restored. An external mesh support[3] was placed around the ascending aorta and root, proximal to the origin of both left and right coronary arteries and secured to the left ventricle with interrupted sutures of 4-0 Ethibond. The mesh was closed anteriorly and secured distally around the origin of the brachiocephalic artery. In view of the aortic regurgitation the transverse dimensions of the external mesh support mesh were scaled during manufacture to be 95% of the aortic root. Postoperative echocardiograms confirmed successful mitral valve

repair and the absence of aortic regurgitation. Follow-up at 11 and 13 months after the surgery was satisfactory.

Discussion

The indications and the management of their severe mitral regurgitation was standard of care for both of these patients with Marfan syndrome. Concomitant root replacement in this scenario is concerning because it exposes patients to an additional operative risk. In addition there are the postoperative consequences of either a mechanical valve with a combined risk from bleeding or thrombosis of 7% per decade or reoperation for failure of the repaired aortic valve presenting a risk of 13% per decade. [6] By halting dilatation while minimising the additional operative risk, concomitant PEARS implantation represents a valuable option in this challenging setting.

PEARS has been available at a limited number of cardiac surgical centres. There are now more than 65 cases with an average follow up of about six years and more than 265 patient years of clinical experience. This provided us with a solution to the clinical problem in two patients with the combination of clinically important mitral regurgitation and a root aneurysm not at usual size criteria for root replacement.

Figure Legends

Figure 1 Historic images related to the first PEARS operation.[7] From left to right, the preoperative MRI, the mesh support on its former, a composite depiction of the position of the mesh, and the MRI in 2014 ten years after placement.

Figure 2. Histology of the mesh/aorta composite. These histological preparations are from the only patient to have died with a mesh in place, more than four years later, of presumed arrhythmia without dissection and with a competent aortic valve.[8] The mesh fibres (yellow arrow) are completely incorporated with collagen fibres running through and around the porous mesh. There are new adventitial blood vessels outside the mesh (green arrow). The aortic media in the unsupported arch has the appearances of Marfan syndrome (central panel) but in the proximal aorta (right) the cardiac pathologists regarded the histological appearances as normal as if there had been healing in the supported segment.

The upper panels show transoesophageal echo images of mid oesophageal aortic valve short axis views before (left side) and after (right side) PEARS implantation in case 1. The Doppler colour flow mapping demonstrated mild central AR; and AR was almost completely abolished after PEARS implantation.

Figure 3. The lower panels show TOE images of mid oesophageal four chamber views before (left side) and after (right side) MV repair in case 2. The Doppler colour flow mapping demonstrated severe MR with jet directed anteriorly. Mitral regurgitation was fully corrected along with MV annular-plasty ring in situ.

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